

Dueling Myxomas: A Rare Finding



Jaime Holbert, DO, MS, Bret Rogers, MD, and Stuart Bresee, MD, *Knoxville, Tennessee*

INTRODUCTION

Echocardiography serves as a cornerstone in the evaluation of ischemic stroke, as cardioembolic events represent 15% of causes.^{1,2} The most common cardioembolic events are caused either by thrombi, often seen in the left atrial appendage in atrial fibrillation, or in association with cardiac masses, such as tumors or vegetations. Primary cardiac tumors are rare and although most often benign are associated with high embolic potential.^{3,4} Myxomas are the most common primary tumors of the heart, most commonly located in the left atrium.³⁻⁸ The discovery of multiple myxomas is a rare finding. We report a case of two left atrial myxomas diagnosed on transesophageal echocardiography (TEE).

CASE PRESENTATION

A 69-year-old man with a history of ischemic cardiomyopathy and atrial fibrillation (anticoagulated with dabigatran) presented with aphasia, right-sided weakness, and right facial droop. His medical history was significant for coronary artery disease with previous percutaneous coronary intervention to the proximal left anterior descending coronary artery 2 years prior, single-lead implantable cardioverter-defibrillator placement for secondary prevention, hyperlipidemia, diabetes mellitus, and distant history of pulmonary embolism. On arrival, he was normotensive at 130/90 mm Hg. Physical examination was notable for aphasia with mild left-sided hemiparesis and an irregular rhythm with soft systolic murmur. Electrocardiography showed a ventricular paced rhythm with underlying atrial fibrillation. Noncontrast head computed tomography did not demonstrate any pathology, but subsequent computed tomographic angiography demonstrated an acute occlusion of the M2 branch of the middle cerebral artery. Given his history of anticoagulation, he was not a candidate for tissue plasminogen activator, so he underwent successful intra-arterial pharmacologic thrombolysis. His acute stroke was tentatively attributed to improper adherence to anticoagulation in the setting of known atrial fibrillation and elevated risk score, as he reported that he was taking dabigatran only daily as opposed to twice daily.

Transthoracic echocardiography was performed and showed chronic severely reduced systolic function, mild to moderate tricuspid regurgitation, mildly reduced right ventricular systolic function, and a

3.7-cm left atrial mass suggestive of a myxoma, seen in [Figure 1](#). The windows for transthoracic imaging were not ideal, leaving open the question of thrombus versus tumor. Cardiac magnetic resonance imaging was contraindicated because of his implanted defibrillator, so TEE was undertaken.

TEE revealed two left atrial myxomas, seen in [Figure 2](#). [Video 1](#) shows both myxomas in the midesophageal aortic valve short-axis view. Three-dimensional (3D) imaging of the myxoma overlying the orifice is seen in [Video 2](#). [Videos 3](#) and [4](#) demonstrate the attachment points of both myxomas. The first myxoma was located near the interatrial septum was 4 × 3 cm, and the second myxoma was seen as overlying the appendage and measured to be 3 × 2 cm. Clear 3D imaging of the attachment points provided by TEE allowed improved preoperative planning as well as intraoperative localization. [Video 5](#) shows how 3D imaging can more clearly identify the stalk of a myxoma. Given the distinctive characteristic findings of myxoma on TEE, along with the location and embolic potential of the dual myxomas, the patient was evaluated by cardiothoracic surgery. Preoperative left heart catheterization was performed and demonstrated significant in-stent stenosis of the proximal left anterior descending coronary artery. Coronary artery bypass grafting of the proximal left anterior descending coronary artery, surgical resection of the masses, and a maze procedure with pulmonary vein isolation and ligation of the left atrial appendage were recommended.

The patient underwent a successful operation. Surgical images are seen in [Figure 3](#). Histopathology was notable for a soft, tan-pink cut surface with a cystic-appearing mass with areas of disruption. His postoperative course was uneventful, with successful discharge to a skilled nursing facility. Postoperative echocardiography demonstrated successful resection of the myxomas and previously reported severely reduced ejection fraction of 25% to 30%. Follow-up echocardiography at 6 months demonstrated persistently low systolic function with an ejection fraction of 25% to 30% and no recurrence of left atrial masses.

DISCUSSION

Transthoracic echocardiography and TEE are key diagnostic modalities in the management of stroke.¹ Given our patient's contraindication to cardiac magnetic resonance imaging, echocardiography was critical in diagnosis and development of the plan of care. On initial presentation, our patient's acute stroke was tentatively attributed to potential nonadherence to anticoagulation in the setting of known atrial fibrillation and elevated risk score. Transthoracic echocardiography, however, revealed a cardiac mass, with TEE confirming the presence of not one but two left atrial myxomas.

Primary cardiac tumors are rare, with the most common being myxomas, accounting for almost half.^{3-5,8,9} Myxomas are typically pedunculated, gelatinous tumors of stromal cells that originate from multipotent mesenchyme capable of both neural and endothelial differentiation.^{4,6} These tumors vary widely in size and are most often identified in the left atrium.^{3,4,7,9} Clinical manifestations include

From the Department of Medicine, University of Tennessee Medical Center, Knoxville, Tennessee.

Keywords: Multiple cardiac myxomas, Transesophageal echocardiography, Cerebral vascular accident

Conflicts of interest: The authors report no actual or potential conflicts of interest relative to this document.

Copyright 2019 by the American Society of Echocardiography. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

2468-6441

<https://doi.org/10.1016/j.case.2019.04.001>

VIDEO HIGHLIGHTS

Video 1: Two smooth mobile masses consistent with myxomas are visualized in the two-dimensional transesophageal echocardiographic midesophageal aortic valve short-axis view. The myxoma on the *left* is attached to the interatrial septum. The myxoma on the *right* is attached overlying the orifice of the left atrial appendage. The implantable cardioverter-defibrillator wire is visualized traversing the right atrium. The aortic valve is tri-leaflet and opens well.

Video 2: Three-dimensional transesophageal echocardiographic video of the myxoma overlying the orifice of the left atrial appendage. The smooth borders consistent with diagnosis of myxoma are observed.

Video 3: Two-dimensional transesophageal echocardiographic bicaval view of a myxoma with typical insertion on the left atrial side of the interatrial septum. An implantable cardioverter-defibrillator lead is visualized in the right atrium. Part of the left atrial appendage is seen on the *right*.

Video 4: Two-dimensional video of a myxoma rocking at its attachment point above the left atrial appendage. The attachment point is clearly identified. The interatrial septum is visualized on the *left*.

Video 5: Three-dimensional transesophageal echocardiographic video of the myxoma attached to the interatrial septum with visualization of the stalk.

View the video content online at www.cvcasejournal.com.

cardioembolic events, as well as constitutional symptoms such as weight loss and fever and laboratory abnormalities such as anemia and elevated C-reactive protein. There is a predilection for women on the basis of several case series. The tumors most commonly present in solitary.^{4,6} The presence of two primary cardiac myxomas is a rare finding, most often seen in patients with Carney complex.

Carney complex, or Carney syndrome, is a rare autosomal-dominant endocrine disorder characterized by multiple benign tumors, such as myxomas, as well as changes in skin pigmentation (lentiginos and blue nevi).^{5,8} Patients with Carney complex are usually young men and have at least two criteria to make the diagnosis, including multicentric tumors and spotty skin pigmentation. Our patient did not have any evidence of pigmented skin lesions, nor did he have evidence of any other endocrine tumors to suggest this diagnosis.

Surgical resection is the treatment recommendation on the basis of risk for embolization.⁹ Recurrence of tumor is rare and is most often associated with multicentric tumors.⁵

Additionally, the use of TEE, particularly with 3D imaging, allows precise localization of the attachment point of these tumors, allowing improved operative planning.

CONCLUSION

Echocardiography serves as a cornerstone in the evaluation of cerebral vascular accident. In our patient, proper diagnosis using echocardiography was paramount to appropriate therapy. The patient was not a candidate for additional imaging clarity with magnetic resonance imaging given the presence of a noncompatible implantable cardioverter-defibrillator. TEE revealed that his stroke was likely the result of the myxomas, requiring definitive surgical therapy.

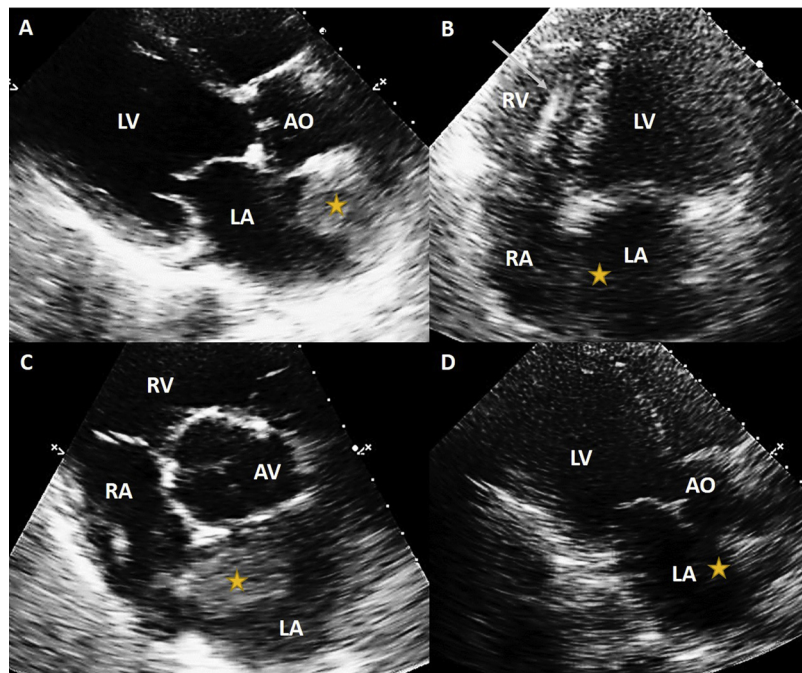


Figure 1 Transthoracic echocardiographic images. The quality of the transthoracic images precludes definitive diagnosis of myxoma and does not provide insight into the presence of multiple myxomas. **(A)** In the parasternal long-axis view, an echogenic mass (*yellow star*) is visualized in the left atrium. **(B)** In the four-chamber view, the left atrial mass is poorly visualized. The implantable cardioverter-defibrillator lead (*gray arrow*) is seen in the right ventricle. **(C)** A smooth echogenic left atrial mass is again visualized in this short-axis view. **(D)** Three-chamber view demonstrating a left atrial mass. AO, Aorta; AV, aortic valve; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

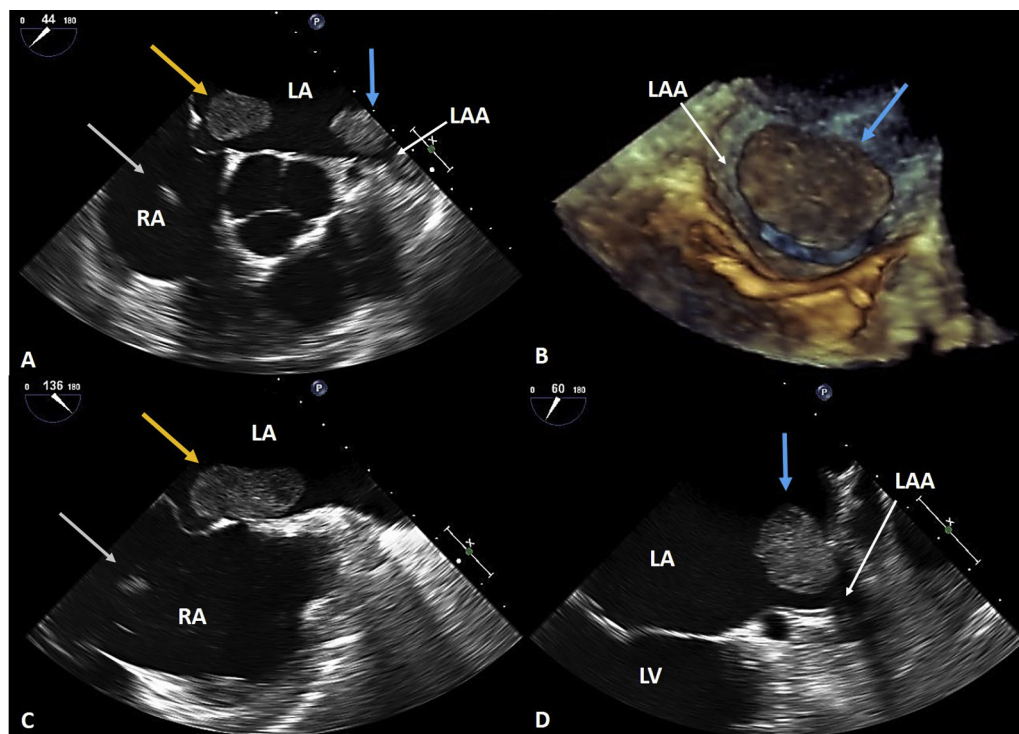


Figure 2 Transesophageal echocardiographic images. **(A)** Two smooth mobile masses consistent with myxomas are visualized in the midesophageal aortic valve short-axis view. The implantable cardioverter-defibrillator wire is visualized traversing the right atrium (*gray arrow*). The corresponding video imaging can be seen in [Video 1](#). **(B)** Three-dimensional imaging of the myxoma (*blue arrow*) overlying the orifice of the left atrial appendage ([Video 2](#)). **(C)** A myxoma (*yellow arrow*) with typical insertion on the left atrial side of the interatrial septum is seen. The defibrillator wire is again visualized ([Video 3](#)). **(D)** Midesophageal view demonstrating a smooth mass with the characteristic stalk of a myxoma attached near the left atrial appendage ([Video 4](#)). LA, Left atrium; LAA, left atrial appendage; LV, left ventricle; RA, right atrium.

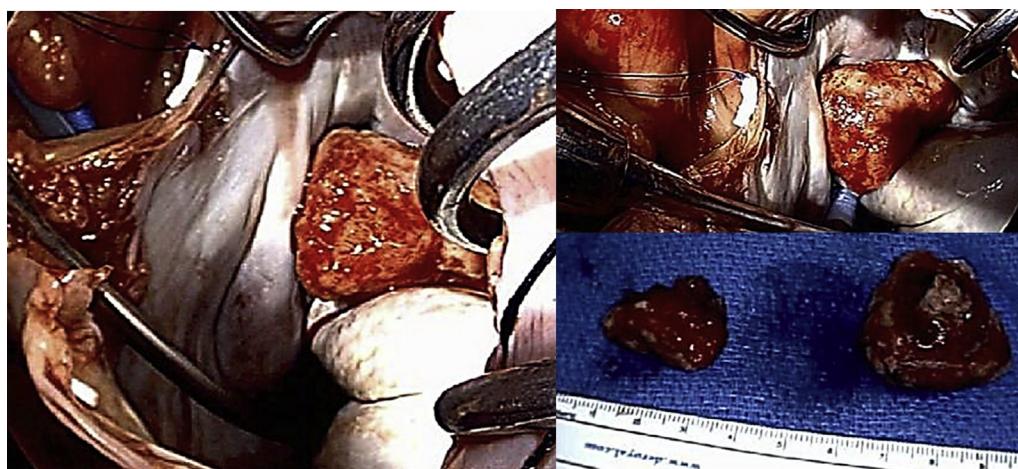


Figure 3 Intraoperative gross images. Gross visualization of the two myxomas after surgical resection.

ACKNOWLEDGMENTS

We thank Erica Massey and Angela Boshears for their assistance in obtaining the echocardiographic images.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2019.04.001>.

REFERENCES

1. Saric M, Armour AC, Arnaout MS, Chaudhry FA, Grimm RA, Kronzon I, et al. Guidelines for the use of echocardiography in the evaluation of a cardiac source of embolism. *J Am Soc Echocardiogr* 2016;29:1-42.
2. Celeste F, Muratori M, Mapelli M, Pepi M. The evolving role and use of echocardiography in the evaluation of cardiac source of embolism. *J Cardiovasc Echogr* 2017;27:33-44.
3. Pontes JC, Silva GV, Benfatti RA, Duarte JJ. Multiple left atrial myxoma: case report. *Rev Bras Cir Cardiovasc* 2011;26:497-9.
4. Lee WC, Huang MP, Fu M. Multiple intracardiac masses: myxoma, thrombus or metastasis: a case report. *J Med Case Rep* 2015;9:179.
5. Carney JA, Gordon H, Carpenter PC, Shenoy BV, Go VL. The complex of myxomas, spotty pigmentation, and endocrine overactivity. *Medicine (Baltimore)* 1985;64:270-83.
6. Okina N, Enzan N, Uchida T, Fukumoto Y. Double cardiac myxomas in the left atrium and ventricle. *Int J Cardiol* 2015;184:364-5.
7. Yuan R, Agha S, Bose S, Blum L, Cleary J, Nair S, et al. Left atrial myxomas presenting with right hand weakness. *Conn Med* 1999;63:261-4.
8. Havrankova E, Stenova E, Olejarova I, Sollarova K, Kinova S. Carney complex with biatrial cardiac myxoma. *Ann Thorac Cardiovasc Surg* 2014;20(suppl):890-2.
9. Wang H, Li Q, Xue M, Zhao P, Cui J. Cardiac myxoma: a rare case series of 3 patients and a literature review. *J Ultrasound Med* 2017;36:2361-6.